



Family Health DataLine

IN THIS ISSUE:

- Birth defects are one of the leading causes of infant mortality and morbidity in the United States.
- Many birth defects are preventable or may respond to early identification through appropriate public health interventions.
- Virtually no data on birth defects exist in Alaska making improvement in public health programs difficult. In response to this, the Alaska Birth Defects Monitoring Program was created.
- Currently medical professionals providing care to children from birth through 5 years of age with one or more of a series of specific birth defects are required to report these children to the Alaska Division of Public Health.

The Alaska Birth Defects Registry

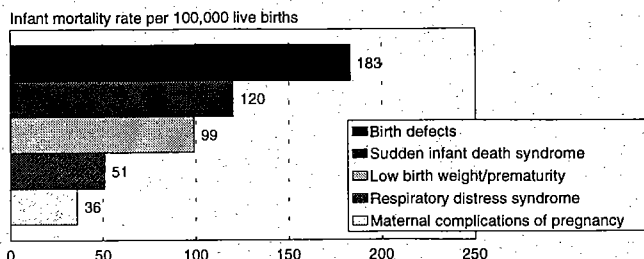
Background

Alaska has joined a growing number of states which have established a birth defects registry. During 1995, the Alaska Department of Health and Social Services (DHSS) promulgated regulations establishing the registry effective as of January 1996. These regulations require all hospitals, physicians, surgeons, and other health care practitioners who have identified a patient from birth through age five (to the sixth birthday) with a birth defect or birth defects to report the patient to the Alaska Division of Public Health within three months of the date of diagnosis or treatment. Birth defects registries have value in improving the health of the nation's children by identifying service needs and improving service delivery to children with birth defects, by identifying areas for targeting interventions designed to prevent birth defects (such as folic acid supplementation to prevent neural tube defects), and by providing information necessary to identify the etiologies of birth defects.

Birth Defects Nationally

According to data from the National Center for Health Statistics, birth defects were the leading cause of infant mortality in the United States during 1992 (Figure 1). The most common major birth defects include cleft lip and palate, cerebral palsy, urinary obstruction, Down syndrome, and congenital heart disease¹ (Table 1). The total lifetime costs associated with children born with a birth defect during 1992 has been recently estimated at more than eight billion dollars. The human and financial costs associated with birth defects dictate that prevention and cost-effective treatment of birth defects have a high public health priority.

Figure 1. Five leading causes of infant mortality; United States, 1992



Purposes of Birth Defects Surveillance

Birth defects surveillance systems are used to conduct investigations, coordinate services, and monitor the outcomes and effects of intervention programs which provide services to children with birth defects. Investigative information available from birth defects registries include baseline prevalence rates and evidence of geographic, temporal, or racial clustering. Additionally, birth defects registries provide the basis for studies investigating etiologic associations, long-term follow-up and outcomes, and program cost-effectiveness.

Birth defects registries have proved their usefulness for epidemiologic investigations on numerous occasions. One of the most notable was the recent documentation that the risk of spina bifida and anencephaly decreased with maternal folic acid intake. Surveillance of these preventable neural tube defects in Alaska will assist in determining whether we are achieving our goals of reducing their prevalence. Another study using data from the National Birth Defects Monitoring Program determined that vinyl chloride was not associated with central nervous system defects. More recently, the Metropolitan Atlanta Congenital Defects Program identified an increasing prevalence of four congenital heart defects, although the reason for this increase remains unknown.

From a programmatic perspective, information from a birth defects registry may be used to address the quality of care provided to children with birth defects. Birth defects surveillance will provide information to assist in the design of prevention and intervention services and improving existing

Table 1. Incidence and cost of various birth defects; United States, 1992.

<u>Condition</u>	<u>Incidence rate (per 10,000 live births)</u>	<u>Total cost (millions)</u>	<u>Cost per new case (thousands)</u>
Cleft lip or palate	17.7	\$697	\$101
Cerebral palsy	12.3	\$2,426	\$503
Congenital heart disease*	10.8	\$1,258	\$345
Down syndrome	10.5	\$1,848	\$451
Urinary obstruction	10.4	\$343	\$84
All birth defects	83.8	\$8,031	\$244

** This includes only truncus arteriosus, single ventricle, transposition of the great arteries, double outlet right ventricle, and tetralogy of Fallot.*

services through better coordination of care.

Recent efforts have identified the etiologic agents of many birth defects including maternal diabetes, the infectious causes of the TORCH syndrome, varicella virus, alcohol, androgenic hormones, isotretinoin, valproic acid, tetracyclines and lack of folic acid. Because their identity is known, the birth defects caused by these agents can be prevented. The Alaska Birth Defects Registry will provide public health officials the data necessary to monitor the effectiveness of current prevention programs and the specific populations toward which new prevention efforts should be directed.

Alaskans are suspected of having unique risk factors for birth defects. Because there is no surveillance system in Alaska to monitor the incidence and prevalence of birth defects as a whole, important epidemiologic and public health factors of birth defects remain unidentified. The birth defects surveillance system that will be implemented in Alaska is expected to form a foundation upon which these issues may be addressed.

Birth Defects Surveillance System Structures in the United States

Birth defects surveillance systems have taken many forms, from the passive acquisition of data through vital statistics to active case ascertainment and reporting requirements. The least expensive method of birth defects surveillance is through collection of data on birth and death certificates. Birth defects surveillance systems which rely on vital statistics data, however, will fail to ascertain the true prevalence of birth defects by as much as tenfold. For example, in Alaska, we have linked birth and death certificate data for children 0 to 19 years of age who died during 1979-92. Of children who died with a congenital anomaly listed as their primary cause of death on the death certificate, 2 of 20 with anencephalus, 6 of 54 with a chromosomal defect, and 0 of 117 with congenital heart disease were identified with these conditions on their birth certificate. Similarly, the Alaska Fetal Alcohol Syndrome Prevention Project found that birth certificates were inadequate to identify children with the syndrome.^{2,3}

Other surveillance systems currently in use by different state health departments have better ascertainment and cost more. A 1997 CDC-compiled directory of state birth defects surveillance programs demonstrates that state health departments utilize, in addition to vital statistics data, hospital discharge records, passive reporting, and active case ascertainment.⁴ The gold standard for current state birth defects surveillance is conducted in California, where data are collected from several sources and staff regularly go to hospitals, genetics clinics, and laboratories to abstract data from logs and medical charts.

Birth defects registries obtain information from multiple sources. Additionally, children with birth defects may have life-long disabilities and present for medical care multiple times. To ensure that each child is counted only once, it is critical to collect unique identifiers. For almost all areas in the United States with birth defects surveillance systems, that means that health care providers and facilities are required to report by name.

Table 2. Reportable ICD-9 Codes for Birth Defects in Alaska Effective January 1996

ICD-9 Code	Condition
237.7	Neurofibromatosis
243	Congenital hypothyroidism
255.2	Adrenogenital disorders
277	Other and unspecified disorders of metabolism
279	Disorders involving the immune mechanism
282	Hereditary hemolytic anemias
284.0	Constitutional aplastic anemia
331	Other cerebral degenerations
334	Spinocerebellar disease
335	Anterior horn cell disease
343	Infantile cerebral palsy
359	Muscular dystrophies and other myopathies
362.74	Pigmentary retinal dystrophy
740	Anencephalus and similar anomalies
741	Spina bifida
742	Other congenital anomalies of nervous system
743	Congenital anomalies of eye
744	Congenital anomalies of ear, face, and neck
745	Bulbus cordis anomalies and anomalies of cardiac septal closure
746	Other congenital anomalies of heart
747	Other congenital anomalies of circulatory system
748	Congenital anomalies of respiratory system
749	Cleft palate and cleft lip
750	Other congenital anomalies of upper alimentary tract
751	Other congenital anomalies of digestive system
752	Congenital anomalies of genital organs
753	Congenital anomalies of urinary system
754	Certain congenital musculoskeletal deformities
755	Other congenital anomalies of limbs
756	Other congenital musculoskeletal anomalies
757	Congenital anomalies of the integument
758	Chromosomal anomalies
759	Other and unspecified congenital anomalies
760	Fetus or newborn affected by maternal conditions which may be unrelated to present pregnancy
760.71	Alcohol—Fetal Alcohol Syndrome

The monitored birth defects may differ by state. Although many states define a birth defect by ICD-9 codes 740-59, others may be more or less exclusive. Some states also include metabolic disorders (such as phenylketonuria or galactosemia), low birth weight infants, genetic diseases such as hemoglobinopathies, and fetal alcohol syndrome (FAS). The age cutoff for surveillance activities varies from the immediate newborn period to 14 years.

The Alaska Birth Defects Registry

The Alaska Birth Defects Registry was designed as a compromise between cost and completeness of data collection. Data are collected from multiple sources including providers, hospitals, laboratories, and programs within the Alaska Department of Health and Social Services. All data collected are stored in a central location with one record kept per child. Information required to be collected include name, race, gender, date of birth, reporting source, and description of the birth defect. Because data are being collected from multiple sources, reporting the name is the most critical component other than the description of the birth defect. All information reported by law to the Alaska Division of Public Health is confidential and may not be disclosed to the public.

As we progress with the Alaska Birth Defects Registry, we hope to work with all providers to develop methods which will reduce the burden of reporting. For our initial reporting, we have developed a reporting form which includes on it the ICD-9 codes which are reportable (Table 2). In addition, the Alaska Division of Public Health

has prepared a booklet titled "Conditions Reportable to Public Health," which is available by contacting Coleen Greenshields at 1-907-269-8000. The booklet provides a summary of the Alaska state regulations relevant to disease reporting and control.

We anticipate analyzing the registry data every year and disseminating results to all contributing groups and to the medical community as a whole. Our goal is to develop a convenient, valid, effective birth defects registry that provides a public health service to Alaska children and communities. To do so, however, we need and seek the assistance of the medical community. For information regarding the registry or to obtain a copy of the reporting form, call Marvin Bailey, Program Manager (1-907-269-3446).

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References

1. CDC. Economic costs of birth defects and cerebral palsy – United States, 1992. MMWR. 1995;44:694-9.
2. CDC. Linking multiple data sources in fetal alcohol syndrome surveillance—Alaska. MMWR. 1993;42; 312-314.
3. CDC. Birth certificates as a source for FAS case ascertainment—Georgia, 1989-1992. MMWR. 1995;44; 251-253.
4. CDC. State Birth Defects Surveillance Programs Directory. National Center for Environmental Health, DBDDD; April 1997.

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